Seizure 1998; **7**: 341–343

Symptomatic nocturnal frontal lobe epilepsy

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INTRODUCTION

Nocturnal frontal lobe epilepsy (NFLE) is a disorder characterised by clusters of brief motor seizures during sleep, often without surface EEG epileptiform abnormalities[1–4]. For these reasons, the prevalence of this disorder has probably been underestimated. Moreover, in the past, many patients with clusters of nocturnal motor attacks have been diagnosed as having nocturnal paroxysmal dystonia[5], paroxysmal awakenings[6] or episodic nocturnal wanderings[7]. At present, except for a case report of a patient with a 'posttraumatic paroxysmal nocturnal hemidystonia'[8], all the patients described in the literature with abnormal motor and/or behavioral nocturnal phenomena had an idiopathic form of the disorder (i.e. absence of aetiological evidence; normal neurological and neuroradiological examinations, and often an unequivocal genetic influence)[1–7]. We studied, by means of nocturnal video-polysomnography, a 39-year-old man with a 14-year history of repeated brief nocturnal motor attacks who had never received a definite diagnosis. His electroclinical picture allowed the diagnosis of nocturnal frontal lobe epilepsy and was indistinguishable from those of typical idiopathic cases. Nevertheless, a magnetic resonance imaging study showed a voluminous cyst arachnoidea compressing the left frontal and temporal lobes. The compression of the frontal lobe, with the possible involvement of the orbitomesial regions, could be the aetiological factors of the nocturnal attacks of our patient.

CASE REPORT

A 39-year-old man, born from nonconsanguineous parents had no family history of epilepsy. The pregnancy, birth and psychomotor development were normal. At age 25 years, he began to show one to three episodes per night with abrupt elevation of head and trunk, fearful expression and dystonic/dyskinetic posturing of the right hand. These episodes, lasting 10–15 seconds, had a high stereotypy in the same night and among the different nights during the 14-year history. The patient was not aware of these episodes, which were described by his mother and then by the wife. Although a psychogenic origin had been postulated, he had never received a definite diagnosis and never been treated for these nocturnal episodes. At age 38 years, he began to complain of difficulty in waking and morning tiredness. He was referred to our Sleep Disorders Center mainly because of these daytime complaints. The neurological examination was normal, as was an EEG performed during wakefulness. The nocturnal video-polysomnography showed four episodes with abrupt elevation of head and trunk, fearful expression and dystonic/dyskinetic posturing of the arms (Figs 1 and 2), three episodes with abrupt elevation of head with fearful expression but with no other phenomena, 22 stereotyped motor attacks with pelvic thrusting, 18 stereotyped motor attacks with scratching the nose and 18 physiological gross body movements. The sleep architecture was the following: sleep latency 43 min, total sleep time 295 min, sleep efficiency 70.8%, stage 1 non-REM 16.7%, stage 2 non-REM 46.3%, stages 3–4 non-REM 9.7%, REM sleep 27.3%, number of stage shifts 92, number of stage shifts/hour of sleep 14.9, total number of motor attacks 47, number of motor attacks/hour of sleep 9.5. The patient did not show either interictal or ictal surface EEG epileptiform abnormalities. Treatment with carbamazepine, 15 mg/kg/day, readily controlled daytime complaints. During the treatment, the reported nocturnal episodes showed a lower frequency (one to three episodes per week), but they did not disappear. After three months of treatment, a second video-polysomnography showed one episode with abrupt elevation of head and trunk, fearful expression and dystonic/dyskinetic posturing of the arms, six episodes with abrupt elevation of head with fearful expression but with no other phenomena, 13 stereotyped

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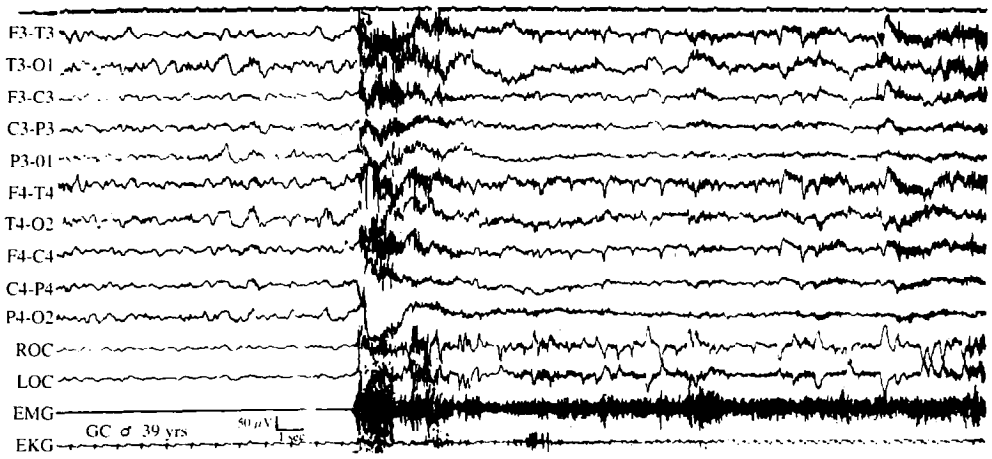

Fig. 1: Polysomnographical recording of an episode, arising from sleep stage 4, with abrupt elevation of head and trunk, fearful expression and dystonic/dyskinetic posturing of the arms. There are no EEG abnormalities. Arrow, onset of the clinical manifestations.

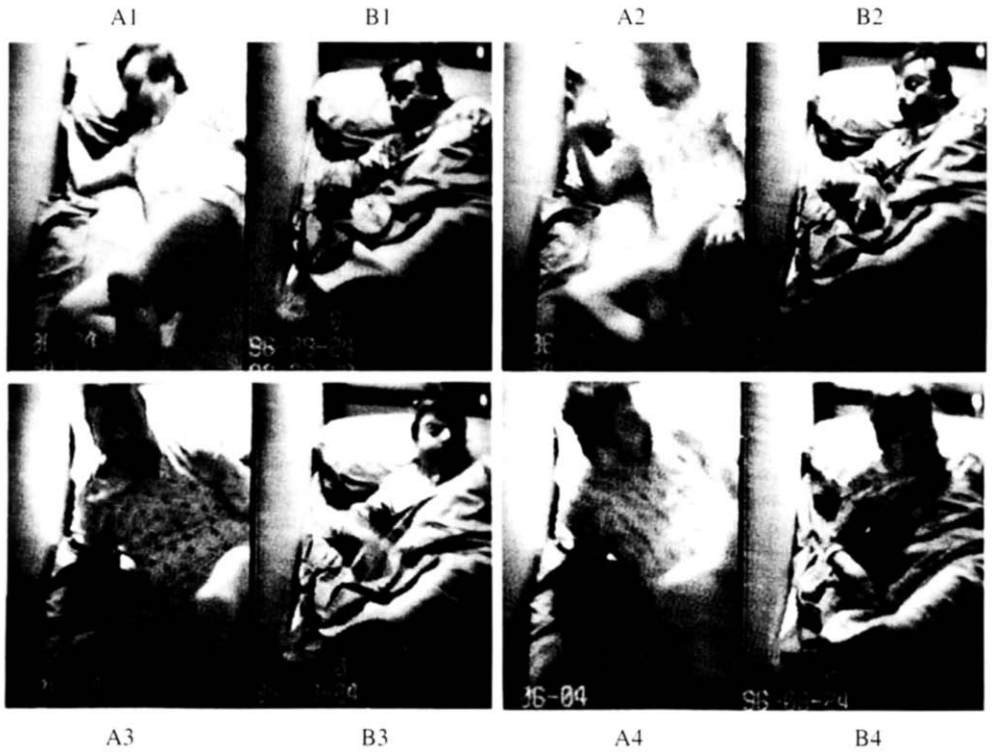

Fig. 2: Photographs of the patient during the episode described in Fig. 1 (A1–4) and during another similar episode in the second polysomnography (B1–4), three months later. Note the stereotypy of the two episode.

motor attacks with pelvic thrusting, four stereotyped motor attacks with scratching the nose and 19 physiological gross body movements. The sleep architecture was the following: sleep latency 21 min, total sleep time 357 min, sleep efficiency 77.9%, stage 1 non-REM 12.7%, stage 2 non-REM 58.0%, stages 3–4 non-REM 12.7%, REM sleep 16.6%, number of stage shifts 104, number of stage shifts/hour of sleep 14.5, total number of motor attacks 24, number of motor attacks/hour of sleep 4.0.

The magnetic resonance imaging showed a voluminous cyst arachnoidea compressing the left frontal and temporal lobes (Fig. 3).

DISCUSSION

The systematic use of nocturnal video-polysomnography has largely improved the diagnostic yield in patients with clusters of nocturnal motor at-

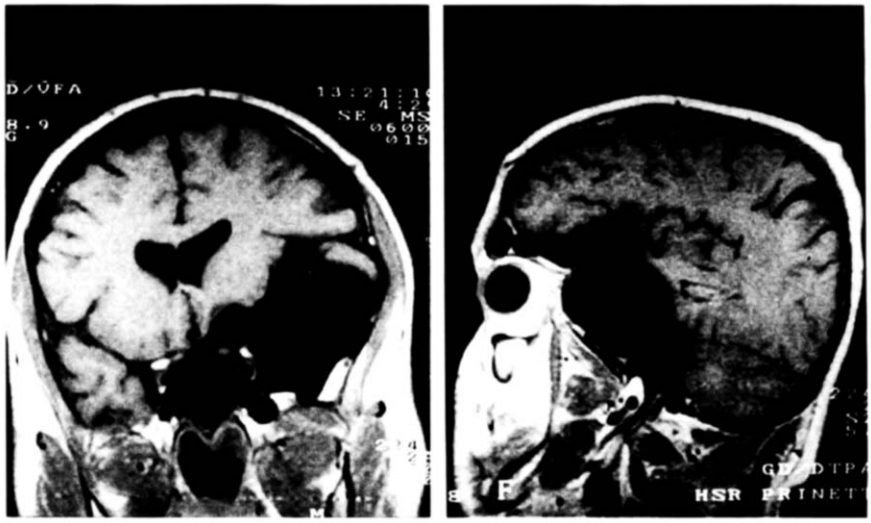

Fig. 3: T1-weighted coronal MRI (left side) and T1-weighted sagittal MRI after gadolinium administration (right side) revealed a voluminous cyst arachnoidea compressing the left frontal and temporal lobes.

tacks. In fact, in the last years, the prevalence of both sporadic and familial NFLE appeared to be higher than previously suggested[2–4]. The electroclinical features and the frequent familial pattern suggest that NFLE is a specific idiopathic localization-related epileptic syndrome, while all the patients previously described in the literature showed neither neurological nor neuroradiological abnormalities[1–5]. Our patient had never received a definite diagnosis probably because of the shortness of the motor attacks and of their absolute nocturnal clustering. Once recorded, he had an electroclinical picture indistinguishable from those of the idiopathic cases of NFLE, but he also showed neuroradiological findings suggesting a symptomatic origin of his disorder. In fact, although the cyst arachnoidea is a benign mass with an origin outside the cerebral lobes, in some cases it can determine a compression of cerebral lobes because of its size[9]. In our patient, the cyst arachnoidea was voluminous enough to cause compression of the left frontal and temporal lobes. In particular, the compression of the frontal lobe and the consequent possible indirect involvement of the orbitomesial regions could be the aetiological factors of these nocturnal motor attacks.

The treatment with carbamazepine reduced most of the nocturnal motor attacks, in accordance with previous papers[2–4]. The great reduction in nocturnal motor-attack-related arousals and, consequently, the reduction in sleep fragmentation, probably produced the disappearance of daytime complaints in this patient.

In conclusion, NFLE is a disorder which is usually idiopathic. Nevertheless, a neuroradiological examination could be important in identifying rare symptomatic cases. Thus, the request for neuroimages for all adults with new onset seizures is probably also valuable for patients with repeated nocturnal motor attacks.

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